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Case Report

Bilateral Choroidal Osteomas in an Elderly Woman: A Case Report

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Keywords

Choroid · Osteoma · Mottled depigmentation · Ultrasound · CT scan

Abstract

Choroidal osteoma is a rare clinical entity of unknown etiology. It is a benign ossifying tumor characterized by mature bone replacing choroid. It typically affects young females, unilaterally. Vision loss occurs mainly due to photoreceptor degeneration secondary to decalcification and/or development of choroidal neovascularization, especially if located near the macular area. We present a case of an old woman with bilateral choroidal osteomas identified incidentally. An 84-year-old Caucasian woman who was asymptomatic, without clinical features suggestive of choroidal osteoma, was referred to our hospital for a follow-up visit. On the fundus examination, both eyes showed a suspected lesion. B-scan ultrasound demonstrated bilateral highly reflective calcified lesions within the choroid, with an evident cone of shadow, suggestive of choroidal osteoma. Further investigations have performed to confirm the diagnosis. Although the literature reports a more common one-sidedness and typical manifestation of choroidal osteoma in the teenage years, our case report refers to bilateral choroidal osteomas in an elderly woman.

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Introduction

Choroidal osteoma is a benign ossifying disorder with the formation of mature cancellous bone in the choroid. The exact etiology is still unknown, and the incidence of the disease is extremely rare [1-3]. It is usually found in women between the 2nd and the 3rd decade

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of life, and the majority of such cases are unilateral and localized close to the optic disc [1]. No risk factors have been identified. Gass et al. [1] first described this entity in 1978. On the fundus examination, it appears as orange-yellow to yellow-white lesions with a distinct margin with blood vessels overlying them. The lesion color depends on the level of overlying retinal pigment epithelium (RPE) depigmentation [4]. They tend to be orange-red in color in the early stages, whereas in later stages, they have a yellowish tint due to RPE depigmentation [2]. The diagnosis is mainly clinical and relies on the appearance of the lesion on the examination of the posterior pole; however, additional diagnostic studies are required to support the diagnosis. Ocular ultrasound and CT, which reveal the bony nature of the tumor, are considered the most diagnostic techniques [3]. The most common causes of visual loss in these patients are due to choroidal neovascularization (CNV) and/or photoreceptor loss and choroidal and RPE atrophy associated with decalcification [4, 5].

Case Report

The publication of this case report was approved by our Institutional Review Board (IRB), and the patient gave a written informed consent to publish her data. The report complied with the principles of the Declaration of Helsinki 1964 and all subsequent versions.

This is a case of an 84-year-old Caucasian woman, asymptomatic, without clinical features suggestive of choroidal osteoma. Her clinical systemic history did not show any renal or para-thyroid disorders, but she was under treatment only for systemic hypertension. Her ocular history did not show any trauma and clinically significant intraocular inflammation.

Bilateral cataract surgery was performed 4 years before, and she was followed for glaucoma in a different center. She was referred to our hospital for a glaucoma second-opinion visit.

Her visual acuity was 20/20 in both eyes with ipermetropic astigmatism correction; intraocular pressure was 12 mm Hg and treated with timolol 0.5 BID, and she was pseudo-phakic in both eyes. On dilated fundus examination, we observed an optic-disc-cup-disc ratio of 0.7 in both eyes and bilateral, yellowish-white lesions with well-demarcated borders located supero-temporally, distanced from the macular area, suggestive of decalcified lesions, one of 8.5 mm × 5 mm × 2 mm in the right eye and the other of 7 mm × 4 mm × 2 mm in the left eye (Fig. 1). No CNV or subretinal hemorrhage was observed.



Fig. 1. Fundus photos showed bilateral yellowish-white lesions with well-demarcated borders located supero-temporally; right eye on the right and left eye on the left.



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Fig. 2. The fundus autofluorescence revealed two bilateral hypo-autofluorescent areas, located outside to the supero-temporal posterior pole edge; right eye on the right and left eye on the left.



Fig. 3. OCT showed a retinal lift with a slight thinning of the retina and a choroidal thickening with some choroidal large vessels. No CNV was detected; right eye up and left eye down.

The fundus autofluorescence revealed two bilateral hypo-autofluorescent areas, located outside to the supero-temporal posterior pole edge, far from the macular area (Fig. 2). The OCT showed a retinal lift with a slight thinning of the retina and a choroidal thickening with some choroidal large vessels (Fig. 3).



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Fig. 4. Irregular hyper-echogenic calcified lesion in the posterior choroid demonstrated by B-scan ultrasound; right eye on the right and left eye on the left.



Fig. 5. Two different CT scans showed an asymmetrical and irregular calcified lesion in the posterior choroid in both supero-temporal choroids; on the right a transversal scan of the orbits, on the left sagittal scan of the orbits.

A visual field test was performed by the Humphrey Field Analyzer II-i (Zeiss, Dublin, CA, USA), program 24-2 SITA Standard, and showed a diffuse slight reduction of the sensitivity in right eye and a superior arcuate defect in the left eye. Because the diagnosis of osteomas must be made by multimodal imaging and ultrasonography, an ultrasound test and a CT scan were prescribed.

A B-scan ultrasound demonstrated an irregular hyper-echogenic calcified lesion in the posterior choroid, focused in both supero-temporal choroids, with acoustic shadowing that gives an appearance of "pseudo-optic nerve" (Fig. 4). The CT scan of her eyes demonstrated an asymmetrical and irregular calcified lesion in the posterior choroid, distanced from the optic disc region, focused in both supero-temporal choroids (Fig. 5).

Discussion

Although the literature reports a more common one-sidedness and typical manifestation of choroidal osteoma in the teenage years, our case report refers to bilateral choroidal osteomas in an elderly woman. The etiology of choroidal osteoma is still unknown. Katz and Gass [6] first described a possible cause of bilateral osteoma, in 1983. They showed a case of



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multiple osteomas developing in association with bilateral pseudotumors of the orbit, which raised the possibility that inflammation might have a part in the cause of secondary ossification [6]. In our case, there were no triggers associated that were reported.

In the largest case series on choroidal osteoma, Shields et al. [4] found that choroidal osteoma showed evidence of growth in 51% of eyes and decalcification in nearly 50% of eyes by 10 years. In their series, decalcification of choroidal osteoma was usually associated with poor vision [4]. Decalcification commonly occurs with overlying RPE alterations and atrophy of the choriocapillaris, both of which could lead to photoreceptor degeneration and poor visual acuity. Shields et al. [7] found that the decalcified portion of osteoma displayed an overlying marked thinning to absent outer retina and photoreceptor layers (100%), compared with the calcified portion with preserved intact outer retina (95%) and intact photoreceptor layer (100%). In our case, there were not any alterations reported, and our patient shows no loss of vision.

Treatment options for choroidal osteoma are limited. Shields et al. [8] also reported photodynamic therapy as a reasonable choice in the case of extrafoveal CNV lesions. However, the authors inserted a proviso at the end of the case report that treatment of subfoveal CNV with photodynamic therapy might result in worse visual acuity due to decalcification and associated RPE loss. More recently, anti-vascular endothelial growth factor drugs have been used off-license to treat CNV secondary to choroidal osteoma with good effect [9].

The early detection of the condition is incidental. Most of the time, patients present with a decreased vision when significant atrophy of retinal layers has already occurred. The correct management involves observation in asymptomatic cases with fundus examination and multimodal imaging at regular intervals, to detect the development of CNV or atrophy of retinal layers, so that loss of vision can be minimized [9].

The differential diagnosis includes choroidal metastasis, sclerochoroidal calcifications, amelanotic choroidal melanoma, amelanotic choroidal nevus, choroidal hemangioma, choroidal granuloma (TB, sarcoid), metastatic calcification, and dysmorphic calcification. In our case, the sclerochoroidal calcification fits the clinical characteristics of the patient because it is generally bilateral, recognized as multifocal yellowish placoid lesions in the supero-temporal and postequatorial area in asymptomatic older white individuals; however, no other ocular or systemic signs were present.

Therefore, in clinics, choroidal osteoma is unique as it affects otherwise healthy eyes as in our patient. The strengths of this case report include a rare disease process involving the choroid bilaterally without significant visual impairment, incidentally, diagnosed in an elderly woman. The limitations of this case report include no follow-up data as our patient did not have previous visits or exams done in our clinic. Our patient will be regularly followed up with six-monthly visits.

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Statement of Ethics

This was a clinical retrospective study, and it was approved by the regional Ethics Committee for publication (CER Liguria: 186/2022). All methods used were carried out according to the criteria set by the Declaration of Helsinki. Written informed consent was



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obtained from the patient for publication of the details of her medical case and any accompanying images.

Conflict of Interest Statement

This research received no specific grant from any funding agency in the public, commercial, or not-for-profit sectors. None of the authors has any proprietary interest in the development or marketing of any products mentioned in this paper.

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Author Contributions

Michele Iester, Cristina Maltese, and Paola Cassottana contributed to the project; Michele Iester, Cristina Maltese, and Paola Cassottana wrote the manuscript; Cristina Maltese, Paola Cassottana, Aldo Vagge, Carlo Enrico Traverso, and Michele Iester revised and approved the final version of the manuscript.

Data Availability Statement

All data generated or analyzed during this study are included in this article. Further inquiries can be directed to the corresponding author.

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